

PRENATAL DIAGNOSIS AND ANTENATAL HISTORY OF TOTAL ANOMALOUS PULMONARY VENOUS RETURN

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SUMMARY

Objective: Total anomalous pulmonary venous return (TAPVR) is traditionally diagnosed by echocardiography or cardiac catheterization postnatally after the appearance of clinical signs. We report a case of TAPVR diagnosed prenatally by targeted echocardiography.

Case Report: A 17-year-old woman was referred at 34 weeks of gestation because of persistent bradycardia. Echocardiography showed atrial disproportion and no direct pulmonary venous return to the left atrium. A female infant was delivered by cesarean section at 38 weeks of gestation. Echocardiography and cardiac catheterization were arranged soon after delivery and TAPVR was confirmed. The infant underwent surgical repair of TAPVR by anastomosis between the left atrium and pulmonary venous confluence as well as surgical ligation of the patent ductus arteriosus at the age of 2 days. The infant died on the 28th postoperative day due to obstruction of pulmonary venous return and respiratory failure.

Conclusion: With advances in sonographic equipment and careful evaluation of cardiac structures, it is possible to diagnose TAPVR prenatally. Pulmonary venous anatomy should be checked during prenatal examination. [*Taiwanese J Obstet Gynecol* 2006;45(3):283–285]

Key Words: prenatal diagnosis, TAPVR, total anomalous pulmonary venous return

Introduction

Total anomalous pulmonary venous return (TAPVR) is an uncommon congenital heart anomaly with abnormal drainage of pulmonary venous blood into the systemic venous system. It may be an isolated lesion or in combination with other cardiac defects. The clinical symptoms and signs are variable and depend on the associated cardiac anomaly. The prognosis of these patients is poor without proper treatment.

Prenatal echocardiography is now widely used in the diagnosis of congenital heart disease [1]. Prenatal diagnosis of cardiac anomaly can offer more comprehensive perinatal management, reduce morbidity and

improve survival [2]. TAPVR is rarely detected prenatally due to its low incidence and difficulty in demonstrating the details of pulmonary venous anatomy [3,4]. We describe a case of TAPVR diagnosed prenatally and the natural course.

Case Report

A 17-year-old, gravida 1, para 0, woman was referred at 34 weeks of gestation because of persistent bradycardia during routine prenatal examination. Echocardiography was performed on GE equipment with a 5 MHz ultrasound probe. Atrial disproportion and failure to demonstrate a direct pulmonary venous connection to the left atrium were found (Figure) and TAPVR was suspected. At 38 weeks of gestation, cesarean section was performed and a female baby was delivered with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively.

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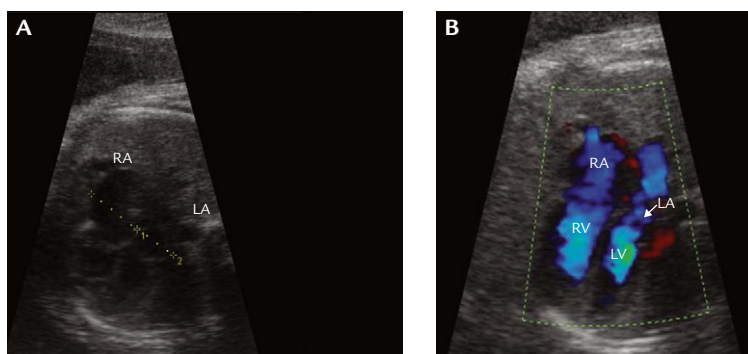


Figure. Fetal echocardiography demonstrates: (A) atrial disproportion; (B) inability to demonstrate a direct pulmonary venous connection to the left atrium. RA=right atrium; LA=left atrium; RV=right ventricle; LV=left ventricle.

Tachypnea and cyanosis were noted soon after birth. Echocardiography showed TAPVR, patent ductus arteriosus (PDA) and atrial septal defect. Triphalangeal thumb of the left hand and partial syndactyly of the right thumb and index finger were also found after birth. Chromosomal study revealed a normal female karyotype (46,XX). Emergent cardiac catheterization was performed the next day and showed mixed type TAPVR with a large right pulmonary vein connecting to the hepatic vein as well as a small left pulmonary vein connecting to the left innominate vein. The infant underwent emergent surgical repair of TAPVR by anastomosis between the left atrium and pulmonary venous confluence and surgical ligation of PDA at the age of 2 days.

The infant was extubated on the 5th postoperative day. After extubation, she showed smooth breathing initially. But tachypnea and subcostal retraction were noted on the 11th postoperative day. Chest X-ray showed increased bilateral pulmonary infiltration at that time. Pneumonia was suspected at first, but her clinical condition did not improve after antibiotic usage. Due to suspected pulmonary venous return obstruction, chest computed tomography (CT) angiography was arranged to delineate postoperative pulmonary venous connection. However, her clinical condition deteriorated and chest CT angiography was postponed. Oxygen saturation became 20–30% on the 24th postoperative day and intubation with mechanical ventilation (high frequency oscillation ventilation) was arranged. But desaturation persisted and the infant died on the 28th postoperative day due to respiratory failure.

Discussion

The incidence of TAPVR has been reported to be 0.008% of live births, with an incidence of approximately 2.2% of patients with congenital heart disease [3]. It could be

an isolated lesion or be associated with various cardiac abnormalities, such as single ventricle, truncus arteriosus, transposition of the great arteries, and cardiac disease with aplenia. TAPVR can be classified by the location of the abnormal drainage as being supracardiac (type I), cardiac (type II), infracardiac (type III) or mixed (type IV), and further designated as with or without obstruction [5].

The clinical symptoms and signs are variable and depend on the pathologic anatomy and the hemodynamic change. TAPVR patients with pulmonary venous obstruction (PVO) usually present with cyanosis and dyspnea, and even sudden death, in the first 2 months, and patients without PVO tend to present with heart failure later. Open heart repair with total correction is necessary in most cases to resolve lethal anomaly. The prognosis of patients without PVO improves after early surgical intervention, whereas patients with PVO still carry high mortality rates [6].

Echocardiography has replaced cardiac catheterization as the standard postnatal diagnostic modality of TAPVR [7,8]. However, only a few papers have described successful prenatal diagnosis of TAPVR [4,9–13]. The prenatal echocardiographic findings of TAPVR we found were atrial disproportion and failure to demonstrate a direct pulmonary venous connection to the left atrium. Atrial disproportion may not be detected until late in pregnancy because of pulmonary blood flow increased in late pregnancy and the presence of atrial septal defect [10]. The pulmonary vein could be identified in two-dimensional echocardiography by interrogation of color and power Doppler.

The natural course of TAPVR is unfavorable because of progressing pulmonary artery hypertension and heart failure. Earlier detection can lead to better perinatal management (delivery in a tertiary care center, earlier delivery, early neonatal catheterization or surgical

interventions). With targeted echocardiography, it is possible to diagnose TAPVR prenatally.

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